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AUTOPSY IN A CASE OF ADIPOSIS DOLOROSA, WITH MICROSCOPICAL EXAMINATION.*

By F. X. Dercum, M.D.,

PROFESSOR OF NERVOUS AND MENTAL DISEASES, JEFFERSON MEDICAL COLLEGE; NEUROLOGIST TO THE PHILADELPHIA HOSPITAL.

The following record is of interest, not only because it is the first autopsy of a case of adiposis dolorosa, with microscopical examination, but especially because it is an autopsy made upon the original case,[1] first published by the writer in 1888. Some two years after this case was described, a similar case was placed upon record by Dr. Frederick P. Henry,[2] and in 1892 the writer grouped these two cases with a third, which he likewise discovered in the wards of the Philadelphia Hospital, and gave to the disease the name of adiposis dolorosa.[3] In 1895, Ewald,[4] described a case which was evidently the same affection, and which he placed in the same category as the cases placed on record by Henry and myself. Subsequently, three cases were described by Spiller,[5] two by Eshner,[6] and one by Giudiceandrea.[7] Dr. Collins[8] also refers to six cases which have been studied by Peterson and Loveland.

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*Read at the twenty-sixth annual meeting of the American Neurological Association, May 1, 2 and 3, 1900.

[1] University Medical Magazine, December, 1888, p. 140.

[2] Journal of Nervous and Mental Disease, March, 1891.

[3] American Journal of the Medical Sciences, November, 1892. Adiposis to be etymologically correct should of course be written adipositas, yet adiposis seems to be sanctioned by general usage.

[4] Ewald, Berliner klinische Wochenschrift, January 21, 1895, page 58.

[5] Medical News, February 26, 1898, p. 268.

[6] The Journal of the American Medical Association, November 12, 1898, p. 1156.

The clinical history of the case here reported has already been published in full. It is briefly summarized as follows:

The patient was a woman, who, when first examined, was 51 years of age. She was a widow, a native of Ireland, and a domestic. The family history presented nothing striking or peculiar. This was also true of her personal history, save that the menopause set in abruptly at thirty-five. Notwithstanding this early cessation of menstruation, she continued to be well until forty-eight or forty-nine years of age. Up to this time she had undergone some general increase in weight, but did not present any of the peculiar symptoms which were noticed later on. A venereal history was denied, but there was probably occasional alcoholic excess.

She first noticed that her arms began to enlarge, and this enlargement was attended by severe pain, shooting and burning in character. It involved both shoulders, the arms, the back and the sides of the chest. It was most marked in the upper arms and back, forming here huge and somewhat pendulous masses. It was elastic, and yet comparatively firm to the touch, and it was impossible to produce pitting. In some situations it felt as though finely lobulated, and in others, especially on the inside of the arms, as though the flesh were filled with bundles of worms. The skin was not thickened; it did not take part in the swelling, and it was not adherent to the subjacent tissues. In addition the swelling was very painful to pressure. Pronounced pressure appeared to be absolutely unbearable. The nerve trunks also were somewhat painful, but this condition was not as marked in them as in the swollen tissue.

The muscles were not involved in the swelling. The affected parts were, however, quite weak. Examined electrically, the muscles of the shoulder and arms yielded a negative result, partly because of the great resistance caused by the intervening tissue. Slight qualitative and quantitative changes were noted in the muscles of the forearms, while in the hands distinct reaction of degeneration was noted in the thenar and hypothenar groups.

Cutaneous sensibility was diminished in various irregular areas over the right arm on both the inner and outer aspects. In the left arm, some impairment of sensation was detected on the outer aspect of the forearm. Sensibility to heat and cold appeared to have been lessened.

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[7] "Un cas d'adipose douloureuse, maladie de Dercum," Revue Neurologique, 7, 1899, p. 877.

[8] "Text-Book on Nervous Diseases," edited by Dercum, 1895, p. 898.

In examining the legs it was found that cutaneous sensibility was distinctly lessened on the right, while showing little or no impairment on the left. There was no alteration of the gait, but both knee-jerks were lost. She complained of a "velvety feel" in the soles of both feet and also in the tips of the fingers.

No enlargement was noted at first in any part of the body, save in the arms and shoulders. There was never any involvement of the face, hands or feet. The face was pale, as were also the mucous membranes. There was, however, a little color in the cheeks, more noticeable at times. Her features were well formed and intelligent. Her hair was dark and fine. Her mind was unimpaired, except that at times she was much abstracted. Sometimes she gave conflicting answers to questions, so that the latter had often to be repeated. The skin seemed dry; sweating appeared to be diminished. There was also some tendency to subnormal temperature during the earlier period of the disease. Her temperature was frequently below 98 deg., and on one occasion as low as 97 deg. After the full evolution of the symptoms, however, this tendency to slightly subnormal temperature was no longer observed.

An examination of the eyes by Dr. de Schweinitz revealed nothing abnormal save some contraction of the visual fields. The other special senses appeared to be somewhat obtunded. An analysis of the urine yielded a negative result. This was also the result of an examination of the blood; there was no leucocytosis.

The patient was under more or less continuous observation at the hospital for a period of eleven years. During this time the neuritic pain, together with increasing enlargement of various parts of the body, formed the striking features of the case. Occasionally small nodules, which had not been discovered before, would make their appearance in various situations. The enlargement gradually involved not only the arms, shoulders and back, but also the abdomen, the hips, the thighs and the legs immediately below the knees. The lower legs, like the forearms, were not affected. The face, hands and feet likewise continued to be free from involvement until the last. On numerous occasions excessive exacerbations of the pains occurred, and not infrequently a simultaneous increase here and there of the swelling would be noted, and the swelling would reveal an increased resistance to the fingers, as though local infiltration or induration had occurred. On several occasions, when the paroxysms of pain were very severe, the patient would be attacked by persistent vomiting, the vomited matter frequently containing blood in small quantities. Vomiting was particularly likely to occur when the paroxysms of pain involved the fatty deposits over the epigastrium. The patient also suffered from frequent attacks of bronchitis, accompanied by dyspnea and blood-stained sputum. Twice she suffered from attacks of herpes zoster, affecting at one time the left arm and left side of the chest, and at another the back. At various times cardiac weakness became marked, and finally led to her death, which occurred March 5, 1899, the diagnosis at the time of death being, in addition to adiposis dolorosa, fatty degeneration of the heart.

The autopsy was held on the next day, and revealed the following conditions:

The body is that of a woman weighing probably three hundred pounds. There is an excessive deposit of fat in the subcutaneous tissue. It is especially marked over the arms, shoulders, back, and to a less extent over the abdomen and thighs. The hair of the pubis is scant, and there is almost none in the axillæ.

In the median line the abdominal fat is 5 cm. in depth. In the epigastric region a small nodule of fat about an inch in diameter is found imbedded in, but separated from, the general subcutaneous fat like an independent lipoma. Several such encapsulated imbedded lipomata were found in various situations; for instance, the back and the right thigh. On opening the abdomen the intestines are well distended with gas; the omentum is filled with fat; the appendix is free, curved and surrounded with fat; the liver extends 2 cm. below the costal margin on the right side; the thorax is well formed; the ribs are well ossified.

Pleural adhesions exist on both sides; the left pleura shows firm adhesions anteriorly and posteriorly. The right pleura shows the same condition as the left.

The parietal layer of the pericardium is covered by a thick layer of fat; the pericardial sac contains 35 cc. of clear straw-colored fluid. The sub-pericardial fat of the heart is very abundant. On opening the right side of the heart, large currant jelly flat clots are found in the right auricle and ventricle. The left side of the heart contains little blood. The heart muscle is friable and of a yellowish color; the wall of the left ventricle measures 2½ cm.; of the right ventricle ½ cm. The aortic valves are slightly thickened; the right auriculo-ventricular opening admits three fingers; the left two. The remaining openings and valves are normal.

The left lung on section shows marked edema; no tubercles are found. The pigmentation is slight. The right lung also shows extensive edema. The spleen measures 17x7½x2½ cm. The capsule is wrinkled; the pulp is soft and dark red in color.

Left kidney is surrounded by a thick layer of fat. The ureter is single. The kidney measures 14x6x3 cm. On the surface and projecting outwards are seen a number of cysts. The capsule strips with difficulty, and the surface is granular. The surface of the kidney is greasy. The cortex is increased, comprising one-half of the kidney structure. The color of the anterior surface is yellowish red. The cortex is lighter than the medullary structure. The right kidney measures 13x7x3 cm., and closely resembles its fellow.

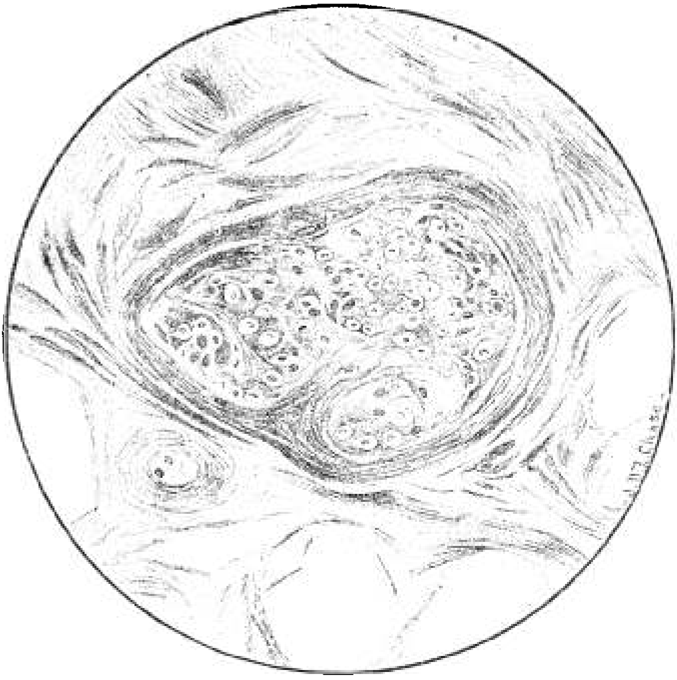

Fig. 1.—Transverse section of a peripheral nerve found in the subcutaneous fat, showing diminution in the number of nerve fibers and proliferation of the connective tissue.

The suprarenals are apparently normal.

The stomach is rather small; walls are not thickened; mucosa shows some post-mortem change.

The pancreas is surrounded by much fat. Section shows no gross change.

The liver measures 29x20x5 cm.; surface is finely granular; edges are irregular and rounded; the knife meets with a good deal of resistance. Color of cut surface is reddish-yellow brown. Interstitial substance well marked. The bile duct is patulous.

The bladder and uterus are normal.

The brain, cord and membranes reveal nothing abnormal.

The thyroid gland is quite small.

The miscroscopical examination included a study of the fatty tissue, the peripheral nerves, the spinal cord and brain, the pituitary body and the thyroid gland.

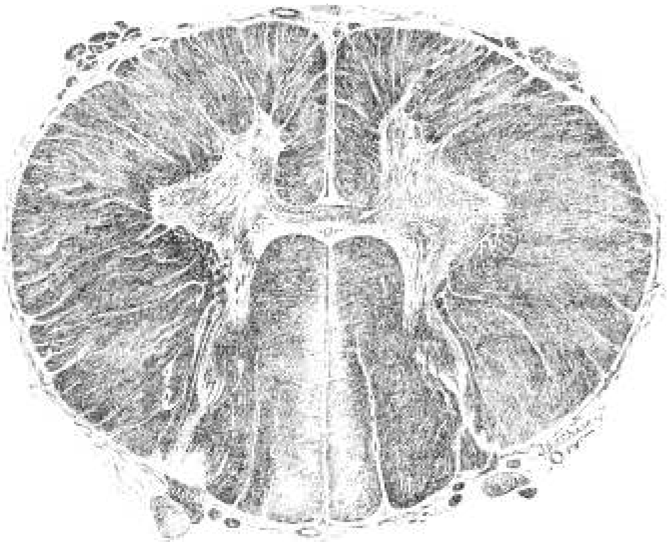

Fig. 2—Section from the upper thoracic region of the spinal cord, showing a moderate degeneration in the columns of Goll.

The fatty tissue in itself presented nothing to distinguish it from ordinary fat. This was not, however, the case with the peripheral nerves found in it. These presented the undoubted evidences of an interstitial neuritis, as will be seen by reference to Figure 1, which shows a transverse section of such a nerve. Here there is seen a marked diminution and atrophy of nerve fibers, together with a marked proliferation of the perineurium and endoneurium. No changes were observed in the larger nerve trunks.

The spinal cord presents some change in the upper thoracic and lower cervical regions, as is seen by reference to Figure 2. There is a slight degeneration in the columns of Goll. This change is most marked in the upper thoracic region. The lumbar cord fails to reveal any changes.

Various portions of the brain examined reveal no change save unusual pigmentation of the cortical cells.

The pituitary body presents nothing abnormal. The acini and the cells composing them present an absolutely normal appearance.

The thyroid gland proved to be exceedingly interesting. Though not weighed at the time of making the autopsy, it was noted as quite small. The microscopical examination of the gland proved it to be abnormal beyond all doubt. (See Figure 3.) In the study of the sections, I received the kind assistance of my friend, Professor Simon Flexner, of the University of Pennsylvania. Very valuable assistance was also obtained from the paper by Dr. Halsted on the effect of the extirpation of portions of the thyroid gland in the dog in Volume I of the Johns Hopkins Hospital Reports.

A study of the sections reveals the gland to be made up of three or four different kinds of secreting tissue. In the first place, there are large acini distended by colloid material. These large acini vary in size, and their contents vary also in density. The larger acini are globular in shape, while some of the smaller ones are elongated or angular in form. The limits of these acini are clearly indicated by blood vessels which occupy their walls. The epithelium is a single layer, which covers uniformly the peripheries of the acini. Contrasted with these there is another kind of secreting tissue, which is very solid, and in which the acini are made out with great difficulty. They consist of cells filling interspaces of the stroma, and the blood vessels supplying these acini can only be made out in exceptional instances. The lumina of these acini when they can be made out are usually very small. There is here a complete absence of colloid material. In other portions acini are observed which are a transition between the more solid

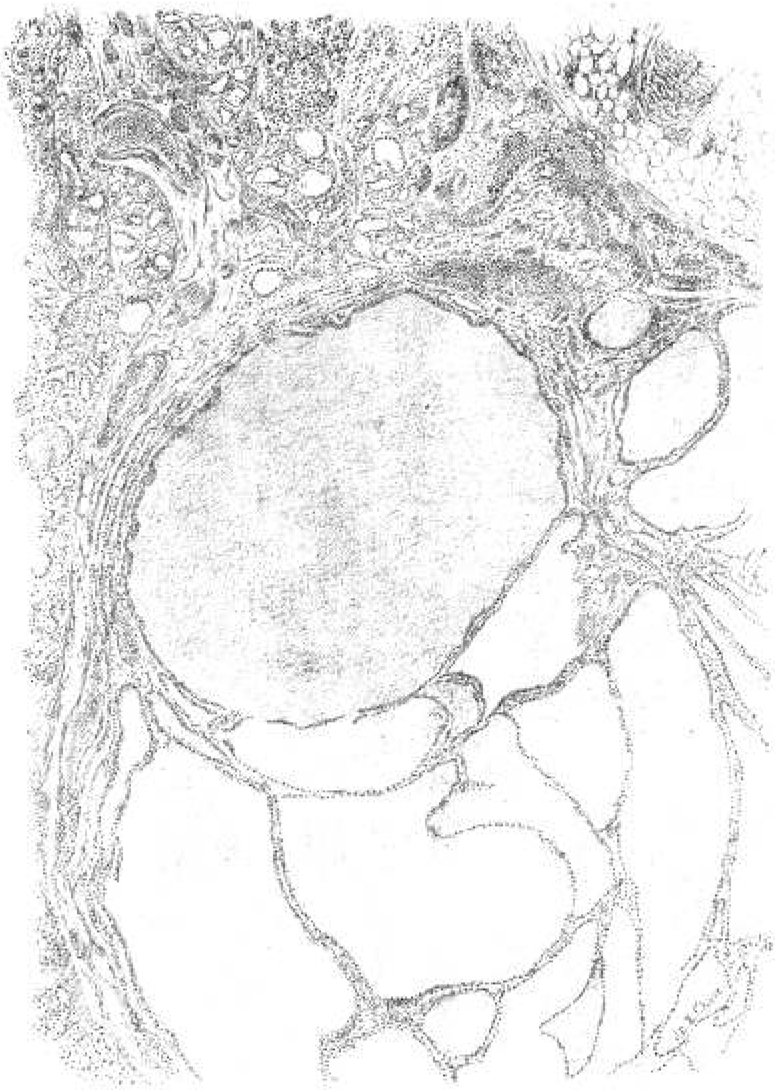

Fig. 3.—Section from one of the lateral lobes of the thyroid gland, showing large acini distended by colloid material. In the central acinus papillary outgrowths of the walls are seen.

nests of cells and the large vesicles which contain the colloid material. In addition, there is a third form of acinus, which is of peculiar interest in that the acini present plications or papillary outgrowths of the walls. These plications or outgrowths project into the lumina of the affected acini, which contain, as a rule, colloid material of lighter staining qualities than the larger vesicles, although not lighter than is contained in some of the smaller vesicles. The epithelium of these last mentioned acini appears at times to be slightly higher than the normal cubical epithelium of the other vesicles. Finally, in some areas, solid masses of cells resembling lymphoid cells are seen, but these are probably young solid acini, like the small acini described above, though the limits of these acini are irregular, because of the absence of preserved blood in the surrounding vessels and of the absence of definite interstitial framework.

The changes observed are indicative in part of hypertrophy. Certainly, this seems to be the only interpretation which can be placed on the numerous small acini which appear to be in process of development. Whether the large acini, distended with more deeply staining colloid material, are to be considered old acini, containing old or altered colloid material, it is, of course, impossible to say, but such an interpretation does not seem improbable. The plications and papillary outgrowths observed in some of the acini are also worthy of comment, in that they evidently represent an attempt to increase the secreting surface of the acini and are again expressive of hypertrophy.

These findings are very surprising, and it is difficult, of course, to frame an explanation. It is not impossible that we have here a hypertrophy which is the direct outcome of a general atrophy of the gland; in other words, a compensatory hypertrophy such as Halsted obtained in the dog after partial extirpation. The gland was small, perhaps sufficiently so to determine compensatory hypertrophy. It is probable, however, that other factors, *e. g.*, qualitative changes of function, also played a rôle in the peculiar symptoms from which this patient suffered. It is not inconceivable that as a result of deranged thyroid action some substance was thrown into the circulation, which at one and the same time prevented the proper oxidation of the hydrocarbons of the food and tissues, and also acted as a cause of neuritis and nerve degeneration. Whatever the explanation, it is interesting to recall the diminished sweating and the occasional slowness of speech and mental irritability; also observed in that other thyroid disease, myxedema.

It is interesting also in this connection to recall that in the two previous autopsies of adiposis dolorosa,[9] the specimens of which, owing to an unfortunate accident, were lost and never microscopically examined, both thyroid glands gave unmistakable evidence of disease. Thus, in one, the thyroid gland was noted as "small, indurated and infiltrated by calcareous matter in both lobes," and in the other as "larger than normal, harder to the feel and much calcified, especially the right lobe."

The writer is indebted to Dr. William G. Spiller for the microscopical work upon the specimens done at the William Pepper Clinical Laboratory.

DISCUSSION.

Dr. J. J. Putnam congratulated Dr. Dercum on his very interesting report. He thought that the subject was of importance from another point of view. The question arises whether the disease of the thyroid gland is the cause of the other changes, or whether both occur together. We should not forget the tendency of the thyroid disease to occur as a sign of systemic degeneration. He knew of an instance where a mother and daughter each had a small goiter. The daughter was congenitally blind. A brother showed infantilism and was also congenitally blind and suffered from progressive muscular dystrophy.

Dr. F. X. Dercum said it was unfortunate that we do not know more about the rôle played by the thyroid gland in simple adiposis. It is well known that in this condition beneficial results are often obtained by the administration of thyroid extract, and in adiposis dolorosa he had seen treatment by thyroid extract followed by very marked improvement, and in one case a complete cure resulted. It is not impossible that the thyroid gland plays a rôle both in painful adiposis and in simple adiposis.

It was interesting that Dr. Burr had noted changes in the pituitary body as well as in the thyroid gland in a case of adiposis dolorosa, though this is hardly surprising, knowing what we do of the compensatory relations which appear to exist between the thyroid gland and the pituitary body.

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[9] American Journal of the Medical Sciences, November, 1892.

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86 AMYOTROPHIE DOUBLE DU TYPE SCAPULO-HUMERAL (Double Amyotrophic Paralysis of the Scapulo-Humeral Type). Georges Guillian (Nouvelle Icon. de la Salpêtrière, Sept., Oct., 1899).

The symptoms in this case developed after traumatism which was unilateral and extraarticular. The case is briefly as follows: A man sixty-three years old, gardener, fell eleven months before in front of a moving carriage. A wheel passed over his right arm. No fracture was found, merely a contusion. Some fifteen days afterward he went back to his work and found that he could raise his right arm with difficulty. There was not any pain and not any swelling. Patient observed on the contrary that his shoulder had grown thinner. This condition grew worse and the left arm became similarly affected.

Examination.—History of heredity or syphilis was absent. The patient presented the typical appearance of a bilateral muscular atrophy of the scapulo-humeral type. Could not raise his arm to a horizontal position, some slight sensory disturbances were present, also some slight pain in the scapular region. Electrical examination showed that faradic excitability was preserved in the trapezius, infraspinatus, rhomboid and deltoid of each side. The contractions were normal. Galvanic excitability was diminished in these muscles. No polar changes observed at all. The tendon reflexes were increased. The further discussion of this case seeks to answer these questions: What is the variety of muscular atrophy present; what is the cause of the condition; what is the anatomical condition, and what is the prognosis? Differential diagnosis had to be made from hysterical paralysis, brachial traumatic paralysis, Erbs' scapulo-humeral myopathy, Aran-Duchenne progressive muscular atrophy, scapulo-humeral type of Vulpian, chronic poliomyelitis, traumatic syringomyelia, pachymeningitis cervicalis hypertrophica and amyotrophic lateral sclerosis. These are all excluded and the condition is believed to be one of bilateral muscular atrophy of peri-articular or articular origin, the so-called reflex type.

The prognosis is not a bad one. Of interest in this case is the bilateral atrophy in spite of the unilateral traumatism. There are a number of such or similar cases reported in literature by Charcot, Raymond and others. The reflex theory of this condition has been proven experimentally by Raymond, who injected nitrate of silver and other irritating substances into the knee joint of animals. Atrophy of the muscles followed. No lesions of the cord have been found in the cases reported. The condition is due to dynamic action and to the irritation of the terminal fillets of the articular nerves.

SCHWAB.